Late-Onset Porokeratotic Eccrine Ostial and Dermal Duct Nevus Associated with Sensory Polyneuropathy and Hyperthyroidism

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This 66-year-old Iranian gentle man had a one-year history of asymptomatic keratotic papules with a linear distribution on the skin of his right palm near the wrist. On histopathologic examination-cornoid lamella-like parakeratotic columns above eccrine ducts were observed. The acrosyringium was also dilated. The diagnosis was porokeratotic eccrine ostial and dermal duct nevus. The late-onset development of the disease in our patient was interesting to us, because it is considered a congenital hamartoma.

Keywords: Acrosyringium • cornoid lamella • eccrine • hamartoma • porokeratosis

Introduction

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) was first reported in 1979 by Marsden et al.1 It was further described by Abell and Read in 1980 in a three-year-old girl.2 Clinically, it is characterized by asymptomatic hyperkeratotic papules with a linear distribution, usually on the extremities.3

Herein, we describe a classic case of late onset PEODDN.

Case Report

A 66-year-old Iranian gentle man was referred to the Dermatology Department of Hazrat-e-Rasoul Hospital by his neurologist because of a lesion on his right palm since one year before. He had been under the treatment of a neurologist for six years due to sensory polyneuropathy. He also had hyperthyroidism.

On the palmar surface of the right hand, there was a patch of yellow-grey punctate keratotic papules arranged in a linear distribution (Figure 1). The lesion was asymptomatic. It was located near the palm-wrist interface, but did not extend to the dorsum of the hand. On examination, there was a collection of 1–2 mm pitted papules. The pits had a keratinous plug that could not be extracted with manual pressure. There was no family history of a similar lesion. Endocrinologic evaluation confirmed hyperthyroidism. In his medical history, he had severe, predominantly sensory polyneuropathy, which had begun six years before. A punch biopsy specimen was obtained from the right palmar skin and stained with hematoxylin-eosin.

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prominent narrow epidermal invagination, with an overlying broad column of parakeratosis; the pattern of cornification resembled a cornoid lamella. The column of parakeratosis extended from a dilated acrosyringium to the surface of the epidermis. At the base of the invagination, there was loss of the granular layer. This feature was limited to the area overlying the acrosyringium (Figure 2).

The acrosyringium was minimally dilated and not filled with any parakeratotic cells (Figure 3). There was also mild chronic inflammatory infiltrations in the superficial dermis. According to the clinical and histopathologic findings the diagnosis of PEODDN was made.

**Discussion**

PEODDN is a rare benign disease which is composed of eccrine hamartoma and cornoid lamellation. Marsden et al. initially described PEODDN in 1979, although they called it “comedone nevus of the palm.” The current nomenclature was coined by Abell and Read in 1980 when they further described this eccrine hamartoma.

The lesions of PEODDN may present clinically either as palmoplantar papules resembling comedones, with keratotic plugs filling the central pits of these lesions, or as keratotic papules and plaques that resemble linear verrucous epidermal nevi on other areas. Lesions may be linear or coalesce into plaques. Two cases with systemic involvement have been described. The lesions most commonly involve the palms and soles unilaterally. Of 19 patients described by Leung et al., three presented with bilateral and 16 with unilateral lesions. Lesions may extend to the dorsum of hands and feet.

PEODDN is generally considered to be congenital, though late-onset cases have been reported. Cases of PEODDN occurring on the upper and lower limbs, forehead, axilla, neck, trunk, and buttocks have also been reported. Sassmannshausen et al. analyzed 24 previously-reported cases and discovered a nearly equal sex ratio (male: female ratio of 12:10, with two patients of unknown sex), absence of family history for PEODDN in all cases, presence of lesions from birth in 15, and involvement of extremities in 23 patients.

The age range for the appearance of PEODDN is from birth to 60 years of age. PEODDN is not associated with other congenital anomalies, though the co-occurrence with linear psoriasis has been noted.

On histologic examination, there is cornoid lamella, which is exclusively associated with eccrine acrosyringia. Cornoid lamella is the histologic hallmark and pathognomonic for PEODDN. The granular layer may be thinned. Vacuolated and dyskeratotic keratinocytes are also typically present within the epidermal invagination. Also, mild lymphocytic perivascular
infiltrates can be observed in the dermis.

In 1992, Bergman et al. proposed that PEODDN is an abnormal keratinizing epidermal invagination transversed by an acrosyringium-like duct rather than by a dilated porokeratotically plugged acrosyringium and dermal duct. This hypothesis was supported by findings of immunohistochemical studies for carcinoembryonic antigen conducted in 1995 and 1996. The differential diagnosis for PEODDN includes nevus comedonicus, linear psoriasis, punctate palmoplantar porokeratosis, linear epidermal nevus, spiny keratoderma, and linear porokeratosis. Linear and punctate porokeratosis presents with cornoid lamellae, involving not only the eccrine gland ostia, but also the opening of hair follicles and the intra-adnexal epidermis. Comedo-like invaginations are absent in porokeratosis.

PEODDN is an asymptomatic disease, but may be a cosmetic issue for some patients. PEODDN is resistant to topical and systemic therapy. As the disease is asymptomatic most of the time, treatment may be reserved for symptomatic patients. Treatment options for PEODDN, which are similar to those for epidermal nevus, include topical steroids under occlusion, ultrapulsed carbon dioxide laser ablation, topical calcipotriol ointment, cryotherapy, electrocautery, and surgical excision. Mazuecos et al. reported a case of PEODDN that showed long-term involution.

In this report, we presented a new case of PEODDN where the palmar skin was the primary site of presentation. The presented case was consistent with the tarda variant of PEODDN, as the onset of lesions was around the age of 65 years. We think that the parakeratotic column seen in PEODDN, although resembles cornoid-lamella, is not true cornoid-lamella, because the parakeratotic cells in this column are very smaller than the cells seen in parakeratosis. We believe that these are not shrunken cells but may be painted keratinous dust. The coexistence of sensory polyneuropathy and hyperthyroidism with PEODDN in this patient was interesting to us. To the best of our knowledge, this combination has not been previously reported, and whether it is a coincidental or true association remains to be uncovered.

References